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Exophytic Variant of Fibrous Dysplasia (Fibrous Dysplasia Protuberans)

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MASAZUMI TSUNEYOSHI, MD

Two cases of an exophytic variant of fibrous dysplasia (fibrous dysplasia protuberans) are reported in which the lesions protruded far beyond the normal bone contour mimicking surface lesions of bone. The first case was an 18-year-old man who had a pedunculated calcified mass of the sixth rib in association with radiologically diagnosed fibrous dysplasia of the skull. The second case was a 33-year-old man who had an exophytic lesion of the proximal tibia. Both of these patients were shown to have benign fibro-osseous lesions consistent with fibrous dysplasia. The intramedullary portions of the host bone adjacent to the exophytic masses also were involved by the fibro-osseous lesions and this fact suggests that the lesions may arise eccentrically in the medullary spaces and mainly grow outwards. It is important to be aware that fibrous dysplasia occasionally presents as an excrescence on the surface of bone. Careful radiographic and histological correlation is required to make a correct diagnosis of this rare variant of fibrous dysplasia. HUM PATHOL 25:1234-1237. Copyright © 1994 by W.B. Saunders Company

Fibrous dysplasia is a benign disorder of the skeletal system characterized by tumor-like intramedullary proliferation of fibro-osseous tissue.¹⁻⁴ It may involve a single bone (monostotic form) or multiple bones (polyostotic form).²⁻⁴ The lesions usually arise within and are confined to the marrow spaces, although expansion or severe deformity of the affected bones is not uncommon in this condition.^{4,5} Fibrous dysplasia is generally not considered radiographically in the differential diagnosis when a lesion extends outside of bone. We report here two unusual cases of fibrous dysplasia of bone forming an exophytic mass, which led to diagnostic problems both radiologically and pathologically.

CASE REPORTS

Case No. 1

An 18-year-old man noticed a painless mass in the left chest wall for 3 years. Plain radiographs and computed tomography (CT) scans showed a parosteal mass with sclerotic fea-

tures of the left sixth rib (Fig 1). An asymptomatic lesion with patchy densities of the left frontal region was found in X-rays and CT scans of the skull during the course of a work-up. Segmental resection of the left sixth rib was performed. A biopsy was not performed on the skull lesion. There has been no evidence of local recurrence of the rib lesion for more than 6 years after the operation.

Pathological Features. On the cut surface the exostotic bony mass, measuring 4 × 2.5 cm, was solid and gray-tan in color (Fig 2). Histologically, the lesion consisted of benign fibro-osseous tissue with broad, irregularly shaped, woven bone trabeculae (Fig 3). Calcified spherules also were scattered in the fibrous component among the bone trabeculae. Spindle cells in the fibrous component were bland and showed no mitotic figures. The fibro-osseous lesion involved the medullary cavity as well, and a diagnosis of fibrous dysplasia was made. In view of the presence of a skull lesion that was radiologically typical of fibrous dysplasia, this case was classified as an example of polyostotic fibrous dysplasia.

Case No. 2

A 33-year-old man noted a painless lump of 2 years' duration on the lateral aspect of the right upper calf. Plain radiographs showed a well circumscribed eccentric lesion with hazy radiodensity in the proximal metaphysis of the tibia. The lesion protruded laterally beyond the normal bone contour producing an exophytic mass with a broad base resembling a sessile osteochondroma. Computed tomography scans showed a well circumscribed protruding lesion on the anterolateral aspect of the proximal tibia with an intramedullary component (Fig 4). A sclerotic border was present on the inner margin of the lesion and the periphery was composed of a continuous shell of cortical bone. The exophytic mass was excised and the intramedullary component was curetted.

Pathological Features. Grossly, the excised exophytic mass, measured 6.5 × 3.0 × 2.0 cm. On the cut surface it was solid and gray-tan in color with a white bony rim along the exterior aspect. Histologically, a fibro-osseous lesion with randomly arranged woven bone trabeculae and bland-looking intervening fibrous tissue was found (Fig 5). Bone trabeculae were not surrounded by osteoblasts; however, osteoclastic resorption was present. At the peripheral edge of the lesion a cap-like zone of sclerotic bone with fibrocartilage was found. Spindle cells in the fibrous area showed no atypia and no mitotic figures were observed. No invasive proliferation pattern into the adjacent soft tissue was observed. A diagnosis of fibrous dysplasia was made.

DISCUSSION

Fibrous dysplasia is a benign fibro-osseous lesion arising in an intramedullary location.¹⁻⁵ Radiologically, fibrous dysplasia usually shows a well defined radiolucent defect with a hazy opacity classically described as "ground glass" in appearance, with or without expansion of the cortex.^{4,6} Eccentric location of fibrous dys-

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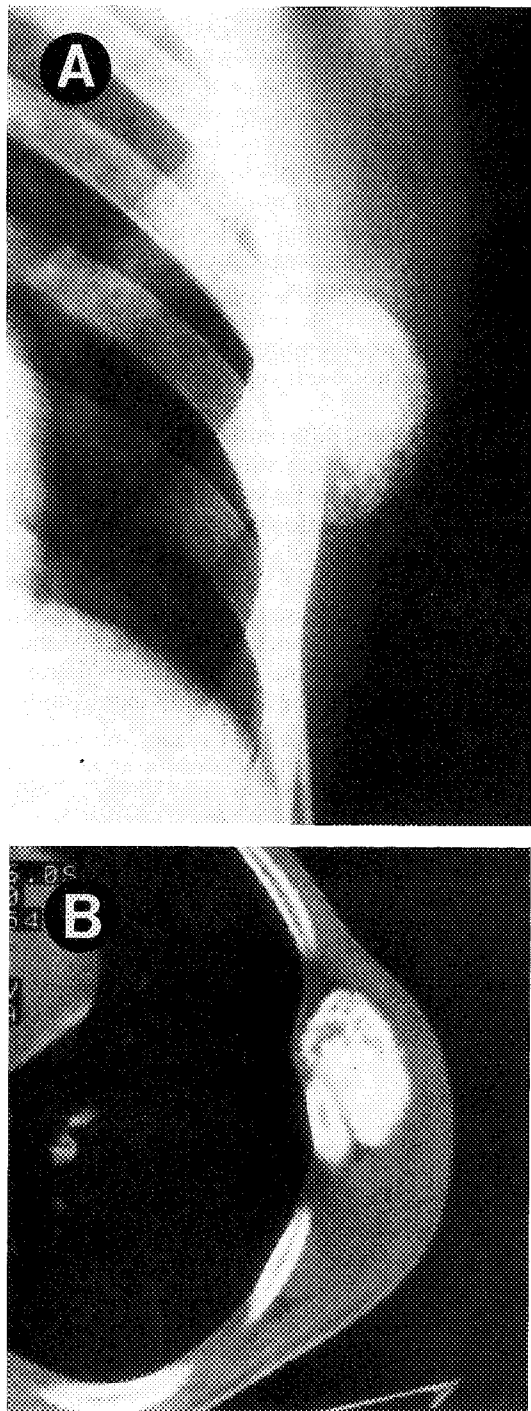


FIGURE 1. (A) Plain radiograph of the right chest shows a calcified pedunculated mass attached to the sixth rib. (B) Axial CT image shows a parosteal mass with calcification.

plasia or localization to the surface of bone is extremely uncommon. Both of our cases were characterized by unusual eccentricity of the fibrous dysplasia lesions and were radiologically interpreted as surface lesions of bone. It is important to be aware that fibrous dysplasia may present as an exophytic mass mimicking an exostosis. We propose the term "exophytic variant of fibrous dysplasia" or "fibrous dysplasia protuberans" for these lesions to emphasize the unusual presentation of this

rare variant of fibrous dysplasia. This lesion is analogous to enchondroma protuberans, a term that has been proposed for eccentric enchondroma.^{7,9}

The exact point of origin of this variant of fibrous dysplasia is unclear; however, both cases presented here had intramedullary involvement. Thus, the lesion may originate eccentrically in the medullary cavity or in the cortex and proliferate mainly outwards rather than within the marrow.

Vigorita et al¹⁰ reported an unusual case of fibrous dysplasia of the middle phalanx of a second toe that appeared as an eccentrically expanding mass with ground glass opacity. This lesion had the radiographic features of a juxtacortical mass. Although these authors addressed the unusual anatomical location for fibrous dysplasia (ie, in a short tubular bone), they did not emphasize the eccentricity of the lesion. We consider their case as another example of fibrous dysplasia protuberans.

Fibrous dysplasia protuberans must be differentiated from benign and malignant surface lesions of bone. Osteochondroma (osteocartilaginous exostosis) is an osteocartilaginous excrescence. The cortex of the stalk of an osteochondroma is continuous with the underlying cortex. Histologically, osteochondroma has a cartilage cap that undergoes growth plate-like enchondral ossification with fatty or hematopoietic marrow being continuous with the host bone marrow. On the other hand, fibrous dysplasia protuberans is basically a fibro-osseous lesion and lacks the composite characteristics of an osteochondroma. Furthermore, the stalk and underlying tissue of an osteochondroma do not have fibro-osseous features. The periphery of a lesion of fibrous dysplasia protuberans may show a fibrocartilaginous cap-like structure, but enchondral ossification is not observed.

Osteoma is the other lesion that must be differentiated from fibrous dysplasia protuberans. Osteoma (ivory exostosis) mostly occurs in the cranium or facial bones, and it rarely involves the long bones.¹¹ In the latter cases the lesion is located parosteally and does not extend into the medullary cavity. These osteomas

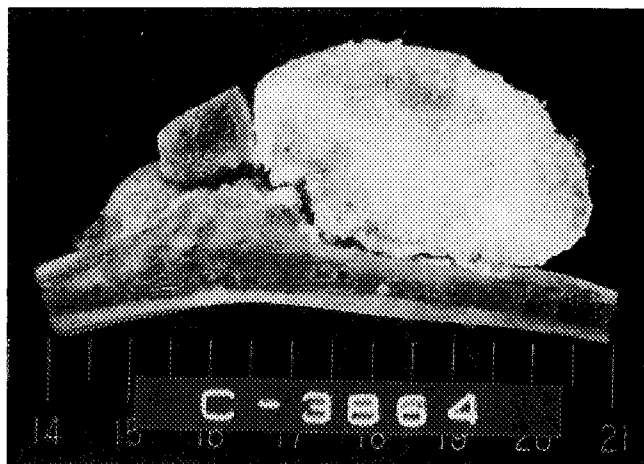


FIGURE 2. Gross specimen showing a protruding bony mass of the rib.

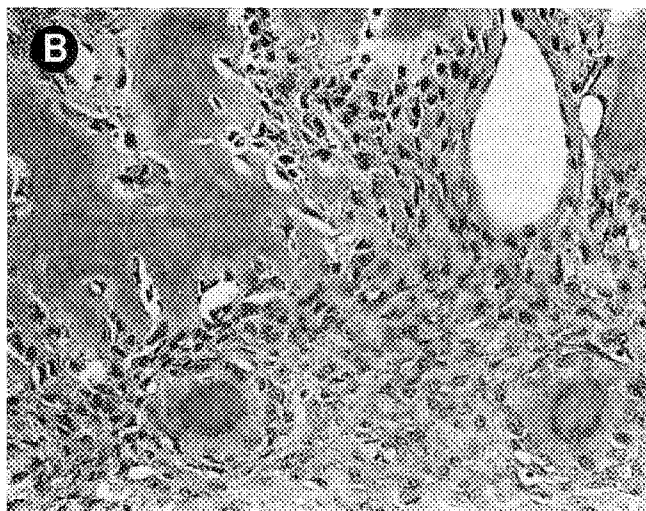
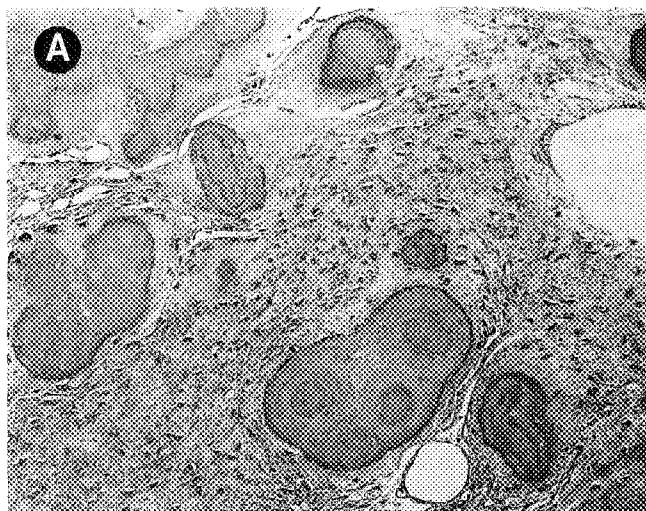


FIGURE 3. Low (A) and high (B) magnification photomicrographs show fibro-osseous lesion with immature bone trabeculae and calcified cementum-like material. (Hematoxylin-eosin; original magnifications: (A), $\times 100$; (B), $\times 200$.)

are frequently associated with tuberous sclerosis or Gardner's syndrome.^{6,12-16} Radiologically, they appear as well circumscribed, dense, bony masses and histologically they consist of mature sclerotic bone without a fibrous stroma.¹⁴⁻¹⁶

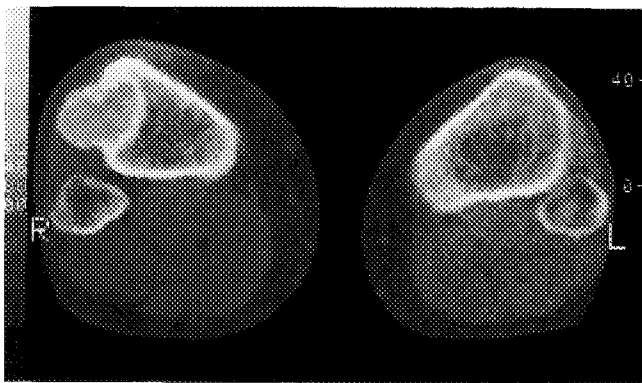


FIGURE 4. Axial CT image of the legs shows a well circumscribed exophytic mass with sclerotic margin of the right tibia.

Parosteal osteosarcoma is a low grade osteosarcoma arising on the outer surface of bone. This tumor roentgenographically shows a radiodense, oval, or spheroid mass attached to the external cortex with a lobulated or irregular margin.¹⁷ In case no. 1 the radiological differential diagnosis included parosteal osteosarcoma, although the rib would be an uncommon site for parosteal osteosarcoma.¹⁸ Histologically, bone trabeculae in parosteal osteosarcoma tend to be arranged in relatively parallel arrays,¹⁹ in contrast to the totally random arrangement in our cases. The presence of calcified cementum-like spherules²⁰ that were observed in case no. 1 has not been reported in parosteal osteosarcoma.

Intraosseous well differentiated (central low grade) osteosarcoma is a distinct variant of osteosarcoma in which the histology may be strikingly similar to that of fibrous dysplasia.²¹ This low grade malignant tumor is composed of spindle cells with only minimal atypia forming osteoid and bone. The differential diagnosis from fibrous dysplasia is made by careful radiographic and histological correlation. Radiologically, the exophytic appearance of fibrous dysplasia protuberans may mimic cortical violation or soft tissue extension of the

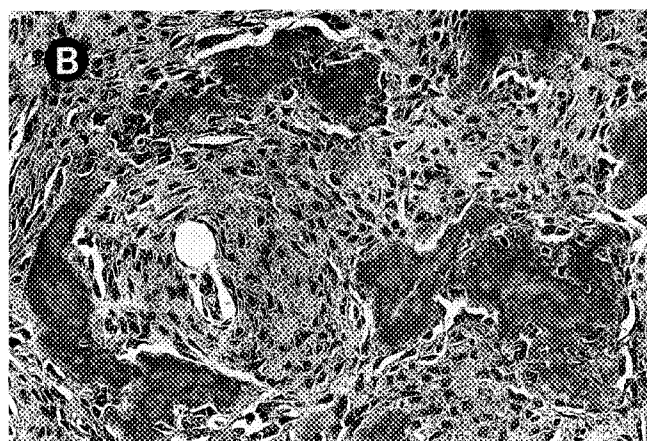
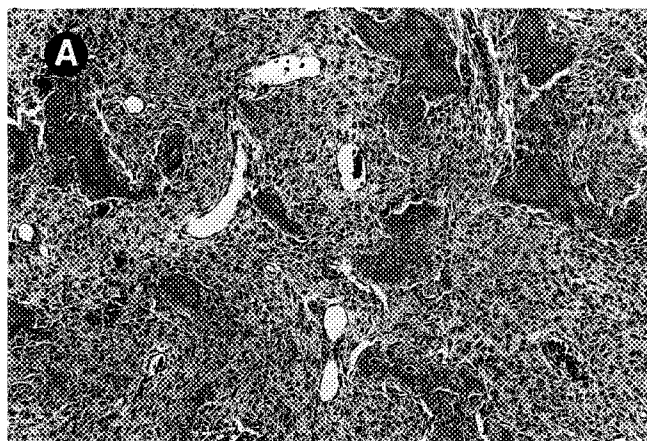


FIGURE 5. (A) Photomicrograph shows benign fibro-osseous lesion with irregularly shaped spicules of nonlamellar bone. (Hematoxylin-eosin; original magnification $\times 100$.) (B) Spindle cells in the fibrous stroma show no atypism. (Hematoxylin-eosin; original magnification $\times 200$.)

intramedullary tumor and may lead to the misinterpretation of aggressive behavior of these lesions. However, detailed evaluation of imaging studies, including CT scans, can help to establish the nonaggressiveness of fibrous dysplasia protuberans.

The exophytic variant of fibrous dysplasia (fibrous dysplasia protuberans) is certainly an uncommon lesion; however, other cases may have gone unrecognized or were misinterpreted as either osteochondromas with fibrotic changes or even low grade osteosarcomas.

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